Transcatheter perforation followed by pulmonary valvuloplasty in neonates with pulmonary atresia and ventricular septal defect

Intérêt du cathétérisme avec perforation et valvuloplastie pulmonaire chez les nouveau-nés atteints d’atrésie pulmonaire avec communication interventriculaire

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Summary

Background. — The classic management of neonates with pulmonary atresia with ventricular septal defect (PAVSD) and moderately hypoplastic pulmonary arteries is usually a systemic to pulmonary artery shunt or ductus arteriosus stenting. We report our experience of transcatheter treatment of PAVSD by perforation followed by balloon dilation of the valve, as it is performed in pulmonary atresia – intact ventricular septum.

KEYWORDS
Interventional catheterization; Pulmonary atresia with ventricular septal defect

\textit{Abbreviations:} PAVSD, pulmonary atresia with ventricular septal defect; PAIVS, pulmonary atresia with intact ventricular septum. 

\textit{Abréviations:} APSO, atrésie pulmonaire à septum interventriculaire ouvert; APS, atrésie pulmonaire à septum interventriculaire intact.

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Patients and method. — Three patients were treated at a mean age and weight of 7.5 days (range 7–8) and 2.9 kg (range 2.5–3.3), respectively. Two newborns were prenatally diagnosed, with micro deletion 22q11 in one case. The three patients had no other pulmonary blood flow support than the ductus arteriosus and were on prostaglandin E1 infusion. The pulmonary atresia was predominantly valvular without significant muscular obstruction. By echocardiography, the mean size of the pulmonary annulus was 6.5 mm (range 6–7). In all cases, the valvular perforation was performed with a 0.014 coronary guidewire, followed by balloon dilation when successfully.

Results. — The procedure succeeded in two cases but failed in the third newborn in whom a long subvalvar muscular stenosis was found at surgery. In the two successful cases, the mean postprocedural transpulmonary doppler gradient was 33.5 mmHg (range 17–50). One patient experienced a femoral venous thrombosis that was successfully treated by heparin and a transient right bundle branch block occurred in another one. In the two successful cases, the prostaglandin E1 infusion was weaned and the surgical repair was performed at 4 and 12 months, respectively.

Conclusion. — In selected cases with PAVSD, perforation followed by balloon dilation of the pulmonary valve offers an interesting alternative to other surgical or transcatheter palliative therapies.

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Introduction

Pulmonary atresia with ventricular septal defect (PAVSD) comprises a broad anatomical spectrum as evidenced by its classification in four stages [1]. Therefore, there are several possible more or less complex management approaches depending on the significance of lesions [1–4]. The most common approach is the surgical systemic to pulmonary artery shunt during the neonatal period, followed by surgical repair when patients are a few months older [1,5–8]. In a recent study, few cases of PAVSD with valve atresia only, a moderately hypoplastic pulmonary tree, confluent pulmonary arteries and no aortopulmonary collaterals, have been treated with perforation followed by valvuloplasty [9]. This technique is similar to that used in the treatment of pulmonary atresia with intact ventricular septum (PAIVS) [10,11] and offers an interesting alternative to the palliative surgical shunt. Nevertheless, the few cases reported in a single centre are not enough to demonstrate the feasibility of this technique [9], insofar as only the successful cases are described. Moreover, indications for this type of catheterization remain poorly defined.

We report our experience with such management in three newborns with PAVSD.
Patients and method

Patients

From January 2001 to June 2007, three newborns with PAVSD were treated with perforation followed by pulmonary valvuloplasty. The mean age was 7.5 days (range 7–8) and the mean weight was 2.9 kilograms (range 2.5–3.3). Pulmonary atresia was purely valvular with a patent infundibulum and confluent but moderately hypoplastic pulmonary arteries. There were no aortopulmonary collateral arteries. The ductus arteriosus was the only source of pulmonary blood flow support in all three cases, maintained with an infusion of prostaglandin E1.

The mean size of the pulmonary annulus was measured at 6.5 mm (range 6–7), while the pulmonary trunk was 6.4 mm (range 6–6.8), the right pulmonary artery was 3.75 mm (range 3.5–4) and the left pulmonary artery was 3.8 mm (range 3.6–4) on the echocardiography.

One of the patients was a carrier of microdeletion 22q11 while the other two had been prenatally diagnosed.

Interventional catheterization

Under general anaesthesia and through femoral vein access, the initial step consisted in a right ventriculogram on lateral view, using a right Judkins 2 or 2.5 curve end-hole coronary catheter (Judkins, Cordis Corporation, NJ, USA). This angiography performed immediately under the valve annulus confirmed the pulmonary atresia and its purely membranous origin (Fig. 1). The 4F right coronary catheter (Judkins, Cordis Corporation, NJ, USA) is positioned just below the valve membranous atresia. Two guidewires

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Figure 1. Left ventriculogram in left anterior oblique view showing the ductus arteriosus dependence of the pulmonary circulation.

Figure 2. Lateral view angiography in the infundibulum: visualisation of Judkins 4F right coronary catheter in contact with the pulmonary valve atresia.

Figure 3. Lateral view angiogram in the infundibulum during perforation of the pulmonary valve by the stiff end of the guidewire.
monary trunk or in the aorta through the ductus arteriosus by using the rigid end of the other guidewire as a slight "rail". The wire that was used to perforate the valve is then removed and another contrast injection in frontal and lateral views is performed to check the position of the soft tip guidewire (Fig. 4). Ballon dilatation of the perforated valve is then performed (Fig. 5).

Results

In one case, perforation of the pulmonary atresia failed due to the narrowness and length of the infundibulum which did not allow correct positioning of the wire. A Blalock-Taussig surgical anastomosis was performed in this patient a few days later. During corrective surgery at 8 months, the infundibulum was described as hypertrophied and particularly narrow.

For the other two cases, the procedure was successful. The main results are reported in Table 1.

One patient experienced a femoral vein thrombosis, which resolved with anticoagulation, and a transitory right bundle branch block was observed following catheterization in the other one. Infusion with PGE1 could be discontinued in the two successful cases with closure of the arterial duct a few days after catheterization.

Biventricular surgical repair was successfully performed at 4 months for one patient and 12 for the other. Preoperative catheterization objectified good development of the pulmonary arteries (Fig. 6).

After a follow-up of 13 and 21 months, neither of these two patients needed any additional surgery or interventional catheterization. The mean hospital length of stay for these children during surgical repair was 10.5 days (range 10–11).

Discussion

Transcatheter perforation of the pulmonary valve using radiofrequency or with a guidewire had already been widely described for PAIVS in the neonatal period [9–11]. The first case was reported in 1991 by Qureshi et al. [10]. Since then, several retrospective studies confirmed its feasibility in 80 to 90% of cases with failures due to the impossibility of perforating the valve [10,11]. Our results confirmed its indication and feasibility for PAVSD where atresia is purely valvular and associated with hypoplasia of the pulmonary arterial tree. Hence, perforation followed by balloon dilation do

<table>
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<th>Table 1</th>
<th>Characteristics of the two successful pulmonary valve perforation.</th>
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<td>Pts</td>
<td>Balloon (diam)/guidewire</td>
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<tr>
<td>1</td>
<td>Tyshak⁶ 6 mm/0.014 BMW⁷</td>
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<td>2</td>
<td>Tyshak⁶ 6 mm/0.014 BMW⁷</td>
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Pts: patients; RBBB: right bundle branch block; diam: diameter; Fem: femoral; Gdt: postcatheterization peak-to-peak gradient; KT: catheterization; min: minutes; SaO₂: transcutaneous oxygen saturation after catheterization and closure of arterial duct; V: vein.

⁶ Tyshak, NuMed, NY, USA.
⁷ BMW, Boston Scientific, Natick, MA, USA.
not lead to congestion due to increased pulmonary blood flow.

However, PAIVS and PAVSD are very different on an embryological and anatomical standpoint. In PAIVS, the right ventricle is hypertrophied but the infundibulum is patent and the pulmonary artery branches are usually well developed [10,11]. PAVSD on the other hand is a conotruncal heart disease occurring much sooner in heart development and due to conal septum malalignment. This anterior and upward malalignment of the conal septum causes obstruction to the right ventricular outflow tract. This obstruction is more often valvular and subvalvular, leading to an extreme form of tetralogy of Fallot [12]. Therefore, the perforation and balloon dilation technique appears to be contraindicated. Only few reports have been published [2,12—15], with the largest study including eight cases over a period of six years [14]. In the case of associated infundibulum narrowing, the procedure was associated with stent implantation [2,13,14]. Our study suggests that this technique may only apply to PAVSD with predominantly valvular atresia, confluent but moderately hypoplastic arteries and ductus arteriosus dependant pulmonary blood flow with no further supply from the aortopulmonary collateral arteries.

The diversity in the management of PAVSD reflects the heterogeneous nature of the lesions [3,4]. When the pulmonary arteries are profoundly hypoplastic with an aortopulmonary collateral dependant pulmonary blood flow, various palliative and curative surgical interventions such as one stage unifocalization of the pulmonary arterial tree are performed [4]. When the pulmonary arterial tree is well developed and dependent on the ductus arteriosus thereby requiring infusion of prostaglandins E1, there are several therapeutic options. Rarely, surgical correction is performed in the newborn [16]. In the majority of the centres, an initial palliative step is performed in the neonate followed by corrective surgery 4 to 12 months later. This palliation may be surgical with a modified Blalock-Taussig anastomosis, which may be complicated by distorsion of the pulmonary arteries, particularly if they are hypoplastic [3,5,6,17]. It may also include interventional catheterization with stent implantation in the arterial duct. Nevertheless, the latter is often technically difficult in PAVSD that is readily associated with a tortuous arterial duct [18]. In addition, the pulmonary blood flow is provided in an asymmetrical and partially retrograde direction, that is less favourable to pulmonary artery growth than an antergrade flow [19].

Historically, this perforation followed by balloon dilation strategy is inspired by dilation of the pulmonary valve in the tetralogy of Fallot suggested about twenty years ago. While this remains a controversial procedure, encouraging results have been published. Yet, indications for such procedure remain limited as this technique concerned the rare patients with a predominantly valvular obstruction. Moreover, the development of successful surgical repair for tetralogy of Fallot earlier in life further limit the indications for balloon dilation of the pulmonary valve [20]. Similarly to balloon dilation, perforation followed by pulmonary valvuloplasty is an interesting alternative in rare cases of PAVSD with moderately hypoplastic but confluent pulmonary arteries, since an antergrade and pulsatile flow offers a better growth potential for the pulmonary arteries [2,12—14]. The induced pulmonary insufficiency also contributes to their development. In addition, this treatment enables an easier access to the pulmonary arterial tree if further diagnostic or interventional catheterization is indicated, for example, in the case of branch pulmonary stenosis [14]. Finally, surgical repair is performed at an age when its morbidity and mortality are lower [2,12—14]. The assessment and extensive use of this technique are restricted by the low number of patients concerned. Moreover, the experience reported on the perforation of PAIVS revealed that complications occur in approximately 15% of cases, while the most common and dangerous include cardiac perforation responsible for rare deaths [10,11]. During the perforation procedure, the wire may perforate the infundibulum, which most often does not lead to major pericardial effusion [13,14]. However, in the Canadian study, the percentage of right ventricular perforation was close to 50% with haemodynamic instability in one patient [14]. In this setting, the use of biplane system enabling to visualise the perforation through frontal and lateral plan is mandatory. In addition, the use of radiofrequency instead of the stiff tip of a guidewire could decrease the rate of perforation. Finally, vascular complications including femoral thrombosis, blood loss with the need blood transfusions are potentially more common in newborns. All this reinforces the concept that this type of interventional catheterization should only be practised by very experienced teams.

**Conclusion**

Interventional catheterization with perforation and dilation of the pulmonary valve offers an interesting alternative to other palliative therapies for some cases of PAVSD with moderately hypoplastic but confluent pulmonary arteries and a patent infundibulum. It only applies to a small number

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**Figure 6.** Frontal pulmonary angiography at 12 months of age showing good development of the pulmonary arteries before surgical repair.
of patients. Its indication should be discussed according to individual cases and in relation to the benefit/risk ratio compared with surgery, which remains the reference method. In all cases, it must be performed by an experienced team.

Conflict of interests

There is no potential conflict of interest.

References